# Case Report

# Elastofibroma: A Rare Case Report and Review of The Literature

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*Elastofibroma is an uncommon fibroelastic tumor-like lesion which usually presents in a characteristic area between the lower portion of the scapula and the chest wall, lying deep to the latissimus dorsi and rhomboid major muscles. It appears almost exclusively in elderly individuals and is associated with history of repetitive tissue injuries. It has pathognomonic histopathologic findings. Although the lesion has previously been defined as a reactive process, its true etiology remains unknown. Based on clinical manifestations and correlation with imaging studies, a presumptive diagnosis of elastofibroma can be made in order to avoid an unnecessary surgery. Here we report a case of elastofibroma in a typical location and present a review of the literature behind its pathogenesis.*

***Keywords:*** *Elastofibroma, Elastofibromatous change, Elastofibroma-like lesion*

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Elastofibroma is an uncommon slowly growing fibroelastic tumor-like lesion occurring in the soft tissue between the lower portion of the scapula and the chest wall, lying deep to the latissimus dorsi and rhomboid major muscles and usually attached to the periosteum of ribs. It was originally named as elastofibroma dorsi because of its appearance in this location[1]. The lesion is mostly defined as reactive process, however its true etiology remains unknown. Although initial reports of this lesion were site-specific, recent reports have shown geographically different distributions[2-7]. It occurs nearly exclusively in elderly individuals over the age of fifty[8] and is associated with history of repetitive activities. Computed tomography (CT) and magnetic resonance imaging (MRI) with clinical correlation allow a presumptive diagnosis of elastofibroma[9] to avoid an unnecessary surgery[10-14]. The lesion has pathognomonic histopathologic features characterized by eosinophilic beaded or string elastic fibers. In the present report, the authors present a classical case of elastofibroma and a review of the literature to discuss its pathogenesis.

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**Case Report**

A 71-year-old Thai male complained of left shoulder pain for 2 months. His symptom was worse when he raised his shoulder. The pain could not be relieved by analgesic drugs or NSAIDs. He denied any history of trauma and long-standing weight-bearing activities. He had hypertension and dyslipidemia. Physical examination revealed an ill-defined firm mass at the lateral chest wall. The mass was deep-seated and not fixed to the skin. No signs of inflammation were noted. Adjacent lymph nodes were not enlarged. The MRI revealed an oval-shaped mass measuring 8 x 6.5 x 3.5 cm located in the deep posterior lateral chest wall on the patient's left side just inferior to the lower pole of left scapula. T1 and T2-weighted images showed intermediate signal intensity without obvious enhancement after gadolinium administration. There was neither bone nor extracompartmental involvement. The radiologist favored a benign tumor but malignancy could not be completely excluded. The tumor was removed and submitted for pathological examination

Grossly, the lesion was an ill-defined soft

tissue mass, measuring 11 x 6 x 5 cm and comprised of a mixture of fibrous and fatty tissue. No areas of hemorrhage and necrosis were seen (Fig. 1). Microscopically, the mass was composed of paucicellular collagenous tissue with entrapped fatty tissue and interestingly predominant degenerative elastic fibers (Fig. 2). The elastic fibers were fragmented into eosinophilic globules or serrated disks resembling beads or strings (Fig. 3). These fibers were stained dark brown to black by Verhoeff-Van Gieson elastic stain (Fig. 4). The diagnosis of elastofibroma was therefore entertained.

The patient had no complications after the operation and no tumor recurrence occurred during four years of follow-up.

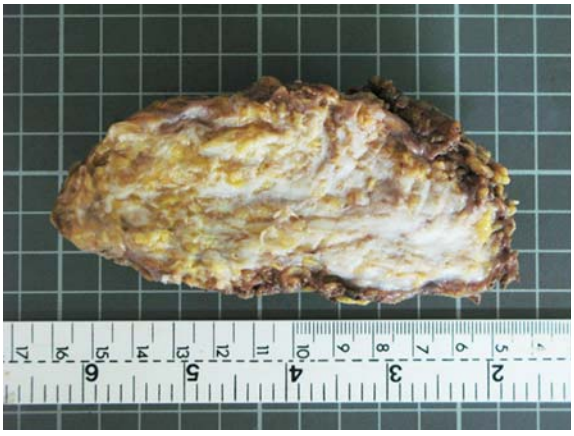

**Fig. 1** Elastofibroma grossly is an ill-defined mass composed of fibrous tissue interspersed by fatty component

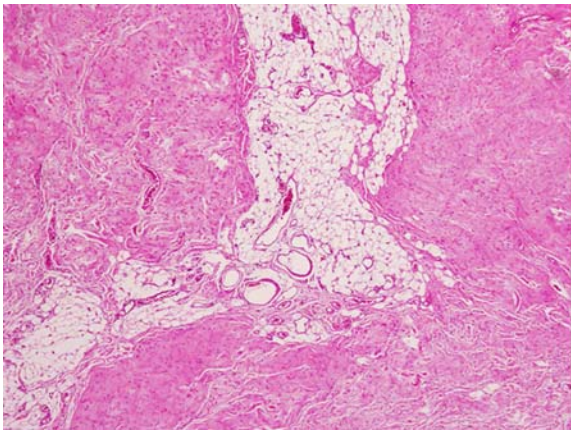

**Fig. 2** Microscopically, elastofibroma contains a mixture of paucicellular fibrous tissue and interlacing fatty tissue (H&E, x200)

## Discussion

Elastofibroma is a benign soft tissue tumor classically found in elderly women and specific to the area between the scapula and the rib cage, deep to the latissimus dorsi and rhomboid major muscles. According to a review of the literature there are regularly worldwide reported cases of this lesion[12-19] and it was also found incidentally at autopsy[20,1]. The authors therefore propose that elastofibroma is not truely a rare entity, but is instead often overlooked due to its painless or asymptomatic features. Originally, its anatomical location and characteristic radiological features made it a site-specific lesion differing from other soft tissue tumors. However other locations have

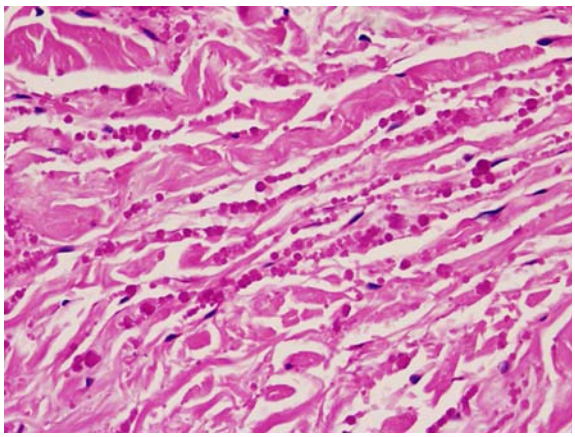

**Fig. 3** Photomicrograph showing the classical fragmented serrated globules or disks resembling beads or strings of elastic fibers are noted (H&E, x600)

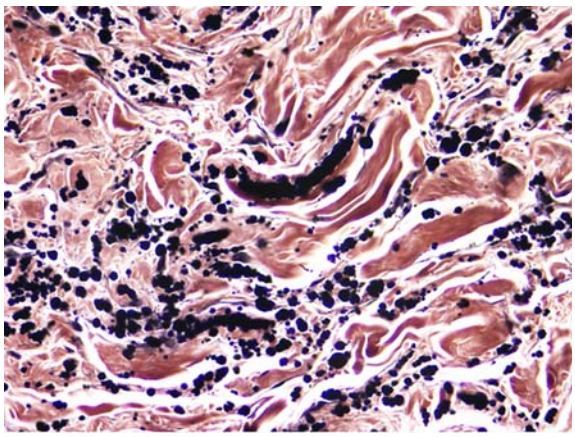

**Fig. 4** Photomicrograph demonstrating typical fragmented elastic fibers stained dark brown to black (Verhoeff-van Gieson staining x600)

been reported in later series such as in the neck[2], stomach[3,4], oral cavity[5,6] and shoulder joint[7]. Even though a right-sided predominance has been previously described, some reported cases have shown bilateral involvement of typical scapular regions[21-23] and multiple elastofibromas[24,25] have been detected as well. The authors question whether it truly has a restricted location and suggest that it is actually a lesion of nonspecific location. For the reason that the scapular region is a predilection area for the friction stress of scapula and chest wall and almost elastofibroma is asymptomatic, the presentation as a scapular mass which can be easily detectable by the patients leads this region to most frequent location.

Most cases of elastofibroma are mainly seen in elderly individuals with a history of manual labor or activities of daily living associated with farm-work and housework[13,18]. Direct mechanical stress on elastic tissue may be an important cause of degeneration of elastic fibers and reactive increase of fibrous tissue. Although there have been rare reported cases of young patients who were athletes and likely to have sports-related traumas[26]. There have also been case series of mimicking lesions suggestive of inflammatory or healing processes in the oral cavity or gastrointestinal tract and which were also diagnosed as elastofibromatous change/elastosis/hyperelastosis[6,28]. These series suggest evidence of a histopathogenesis involving both reactive and degenerative changes. However, not all reported cases could disclose definite evidence of an explanatorytrauma or injury, including our particular case report[3,27]. Perhaps the traumas might cause extremely minor injury and were not concerned by the patients or physicians.

On the other hand, significant chromosomal instability was found in elastofibroma[17,29-32]. These evidences support a possible neoplastic origin. In addition, there have been case reports of family members with bilateral elastofibromas, suggesting a genetic etiology[29]. A large series of Japanese patients has detected hereditaryand constitutional influences for elastofibromas[8]. Some researchers suggested that abnormal elastic fibers are immature fibers produced by activated fibroblasts rather than degenerated fibers[33,34]. Kuroda et al have proposed that fibroblasts may produce abnormal elastic fibers and collagen fibers through the secretion of TGF-β[35]. However, a study of Keita et al disagreed with this theory because they could not demonstrate significant different levels of TGF-β between elastofibroma and control[4] but they instead suggested the vascular-concentric development and active neovascularization of the elastofibroma. Additional studies in large series are needed to define the presence of common genomic alterations to characterize biological significance of these genomic abnormalities in elastofibroma.

Histopathologically, elastofibroma revealed a pathognomonic feature characterized by fragmentation of the elastic fibers into serrated globules or disks resembling beads or strings. These elastic fibers are highlighted by Verhoeff-van Gieson stain. Based on immunohistochemistry study, the spindle cell component in elastofibroma is stained with a specific antibody for CD34[17,35] but shows negative stainings for smooth muscle markers including alpha-smooth muscle actin, h-caldesmon[35], desmin and neural maker namely S-100[36]. These findings should suggest fibroblastic origin of the lesion.

Elastofibroma displays typical characteristics on imaging studies. On CT scan, it appears iso-dense with hypo-dense strands of fatty component. MRI is a modality of choice for investigation because it has high level of accuracy for diagnosis[22,37,38]. The lesion appears as a low to iso-intensity signal compared to surrounding muscular structures on T1 and T2-weighted sequences with internal strands of fatty signal. Interestingly, elastofibroma results in low grade diffuse F-18 FDG uptake on PET scan[39-41]. These radiographic findings, in combination with specific clinical features including advanced patient age, female gender and specific scapular location, can be used to diagnose elastofibroma prior to removal. Because of its slow-growing low-grade lesion without report of malignant transformation, observation is the best choice of management in asymptomatic cases in order to avoid unnecessary surgery. Complete excision is a treatment of choice only in cases with certain symptoms or functional disabilities[10-14]. Elastofibroma has very good prognosis. To the best of our knowledge, malignant transformation of this lesion has not been reported in the literature. There is also no evidence of recurrence after complete resection[11,16,42]. Tissue biopsy should be performed for cases presenting in atypical locations or suggestive of other soft tissue tumors especially in cases that sarcomas cannot be excluded.

In summary, the pathogenesis of elastofibroma is still controversial. Mechanisms involving reactive, degenerative and true neoplastic processes have been proposed. Clinical manifestations in combination with imaging features can lead to definitive diagnosis and prevent unnecessary sugery.

**Acknowledgement**

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**Potential conflicts of interest**

None.

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## *Elastofibroma: รายงานกรณีศึกษา และทบทวนวรรณกรรม*

***จุฑาทิพย์ คินทรักษ์, บัญชา ชื่นชูจิตต์***

*Elastofibroma เป็นก้อนคล้ายเนื้องอก (tumor-like lesion) ประกอบด้วย fibrous และ elastic tissue พบบ่อยที่บริเวณระหว่างด้านล่างของกระดูกสะบัก และผนังทรวงอก โดยอยู่ลึกลงไปจากกล้ามเนื้อ latissimus dorsi และ rhomboid major มักพบในผู้สูงอายุที่มีประวัติการบาดเจ็บของเนื้อเยื่อซ้ำๆ ลักษณะทางพยาธิวิทยา มีความจำเพาะและให้การวินิจฉัยได้ แม้เดิมเชื่อว่าเป็นพยาธิสภาพที่เกิดจากการปฏิกิริยาตอบสนองต่อ การบาดเจ็บของเนื้อเยื่อ แต่ปัจจุบันยังคงไม่ทราบสาเหตุที่แท้จริง Elastofibroma สามารถวินิจฉัยได้ ทางคลินิกโดยอาศัยประวัติ ตรวจร่างกาย และการส่งตรวจ imaging study เพื่อหลีกเลี่ยงการทำผ่าตัดที่ไม่จำเป็น ผู้เขียนรายงานกรณีศึกษา elastofibroma และทบทวนวรรณกรรมเกี่ยวกับพยาธิกำเนิด*

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